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ARRESTED CEREBRAL DEVELOPMENT,

WITH

SPECIAL REFERENCE TO ITS CORTICAL
PATHOLOGY.

BY

B. SACHS, M.D.,

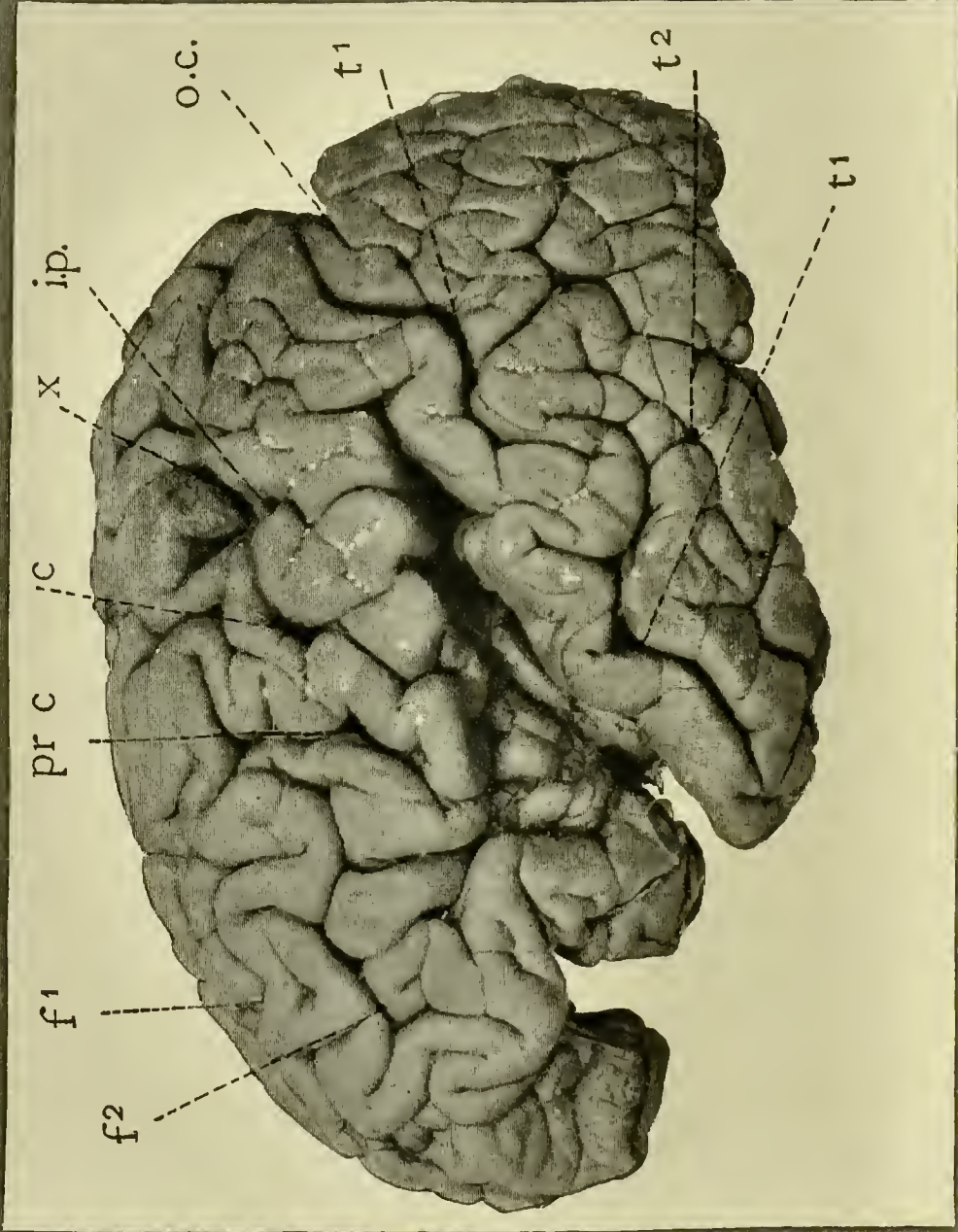
INSTRUCTOR IN MENTAL AND NERVOUS DISEASES AT N. Y. POLYCLINIC; CLINICAL ASSISTANT
TO THE CHAIR OF NERVOUS DISEASES, COLL. OF PHYS. AND SURGEONS, NEW YORK.

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PLATE I.



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ON ARRESTED CEREBRAL DEVELOPMENT,
WITH SPECIAL REFERENCE TO ITS COR-
TICAL PATHOLOGY.¹

OUR knowledge of the pathological substratum of the various forms of mental derangement is still very imperfect. In the majority of cases, there may be no marked changes in the structure of the brain; or, if there be any changes at all, they are entirely beyond our ken, and cannot be made out by our present methods of investigation. As mental pathology is in its infancy, it is but natural that we should first seek for structural changes in those conditions in which the departure from the normal is greatest, in which the mind is disturbed, as a whole, and not merely with reference to a single part or faculty; though I shall at once declare my belief that derangement of a part of the mind means disorganization, more or less complete, of the entire mental mechanism.

While we are even now in possession of many facts concerning the morbid structural changes in dementia paralytica, changes that accompany the complete *dissolution* of a fully developed, and once normal mind, we have busied ourselves but little with the morbid changes that often

¹Read before the American Neurol. Assoc., July, 1887.

affect the brain, and consequently the mind also, when both are yet undergoing the process of *evolution*. These cases of retarded development, of idiocy, of mental imbecility, call them what you will, seem to me to possess a deep pathological and physiological interest. From the pathological changes found in these cases of extreme mental defects, we are entitled to draw an inference regarding the normal function of those nervous elements here found deficient, and we may well argue with regard to such broad facts as an absolute lack of mentality, although it may be a long time to come before we shall be able to explain the morbid mechanism underlying fixed delusions, hallucinations, and the like, or to state exactly what the structural changes are in paranoia, in circular insanity, and in other grave mental troubles.

The condition which I have the privilege of discussing before you to-day represents not only such changes as come about in the process of evolution, but represents changes of the earliest period of infantile development.

Much has been written upon idiocy and allied conditions from the clinical point of view, but pathological and pathological-anatomical observations are surprisingly few and far between. And those who give gross morbid changes fail to refer to the histological changes either in the cortex or other parts. Thus Bourneville,¹ who has made an excellent contribution to the study of this subject, refers in only one of five cases to changes in the cortex. Brückner² has given the most detailed account of the histological changes in the cortex with which I am acquainted. His was a case of what is known as tuberous sclerosis of the cortex, and concerned a patient 22 years of age. The pathological changes underlying these conditions of idiocy are undoubtedly as varied as the clinical manifestations themselves; for the present we designate these affections by broad clinical terms; later on we may

¹ Bourneville ("Sclerose tubereuse des circonvolutions cerebrales"). Arch. de Neurologie, vol. i.

² Brückner, "Ueber multiple tuberöse Sklerose der Hirnrinde" (Arch. f. Psych., vol. xii.).

be able to differentiate between them, and to give to each condition its proper pathological designation. Looked at in this way, the title of my paper is altogether too comprehensive. The changes which I have to report upon to-day are a few of the many changes which may give rise to similar clinical mental phenomena.

Before presenting the history of the case, I must acknowledge my indebtedness to Dr. I. Adler, of New York, through whose kindness I was enabled to observe the case closely, and with whom I shared the responsibilities of treatment; and to my friend, Dr. van Gieson, who was kind enough to supply me with normal material of the same age for comparison, and who, during an unexpected absence from the city, assisted me in the work of cutting and staining.

The following is the history of the case: The little girl S., who was but two years old at time of death, was the first-born of young and healthy parents. In the families of both parents insanity is not unknown; on the mother's side there is a strong hereditary predisposition to mental disease, and several near relatives of the father have developed various forms of insanity within recent years. During the fifth month of pregnancy, the mother was thrown out of her carriage, but did not sustain any serious injuries; the child was born at full term, and appeared to be a healthy child in every respect; its body and head were well proportioned, its features beautifully regular. Nothing abnormal was noticed until the age of two to three months, when the parents observed that the child was much more listless than children of that age are apt to be; that it took no notice of anything, and that its eyes rolled about curiously (there was evident nystagmus). Allowing for some very slight vacillations, the child remained in practically the same condition up to time of death. The condition was characterized as follows: The child would ordinarily lie upon its back, and was never able to change its position; muscles of head, neck, and back so weak that it was not able either to hold its head straight or to sit upright. It never attempted any volun-

tary movements; movements that were made were in obedience to peripheral stimulation. All the muscles were extremely flaccid; all reacted perfectly to both forms of current. The child would close its hand upon the finger of the examining person, but objects placed in its hands were quickly dropped. The child as it grew older gave no signs of increasing mental vigor. It could not be made to play with any toy, did not recognize people's voices, and showed no preference for any person around it. During the first year of its life, the child was attracted by the light, and would move its eyes, following objects drawn across its field of vision; but later on absolute blindness set in.

Dr. Knapp, who made several ophthalmoscopic examinations of this case, reported the following unusual condition, at the seventeenth meeting of the Heidelberg Ophthalmological Society. The report may be found in the Proceedings of this meeting. Dr. Knapp there says: "Child two to three months; nystagmus vibratorius; pupils contracted as is usual with children at this age. Media clear, optic nerve discs pale. Fovea centralis, of a cherry red color, was surrounded by an intense grayish-white opacity. This opacity was most distinct in the vicinity of the fovea centralis, and for some little distance around it, but faded away gradually into normal retinal field."—Dr. Knapp at first gave a favorable prognosis, except as regards central vision, more particularly as there appeared to be for some time a slight improvement in vision. He could not then, and is not now ready, to give an explanation of this condition.

But two cases of this sort of retinal changes had thus far been reported, by Magnus and Goldzieher, and neither of these authors has any explanation to offer. Dr. Knapp, in private conversation, hinted at a developmental defect. Unfortunately, the eyes could not be removed after death. Dr. Knapp empowers me to add that "a further examination in May and June, 1886, revealed great changes. Child totally blind, optic nerves completely atrophied (discs as

white as paper, with scarcely a trace of blood-vessels). Macula lutea essentially as before."

By way of anticipation, it may be remarked that numerous longitudinal and vertical sections of both optic nerves were variously stained and examined, but that no morbid changes could be made out. Blindness must, therefore, have been due either to the retinal changes, or to the deficient cortical condition, or to both.

Hearing seemed to be very acute; there was unusual hyperexcitability to auditory and tactile impressions; the slightest touch and every sound were apt to startle the child. The child never had convulsions, not even while teething; no marked rigidities at any time. The child never learned to utter a single sound; if left to itself it would occasionally make a low gurgling noise. Bodily functions normal, excepting the frequent recurrence of bronchial troubles and feebleness of its digestive powers. At the age of one it had a severe attack of diphtheria from which it rallied in the course of a few weeks. The child developed unusually high fever with every disturbance, however slight, of its bodily functions. In the way of treatment nothing was recommended but careful nursing and feeding, tonic treatment with malt and the like; phosphorus was given in small doses for a time, and the peripheral muscles and nerves were alternately galvanized and faradized, more in the hope of exciting cerebral activity in a reflex way than of benefiting the nutrition of the flaccid parts.

There were no distinct evidences of inherited or acquired syphilis and none of rachitis.

During last summer (1886), the child grew steadily weaker, it ceased to take its food properly, its bronchial troubles increased, and finally, pneumonia setting in, it died August, 1886.

Immediately after its death, the child was brought to the city, and yet twenty-nine hours had elapsed before the autopsy could be made.

Autopsy.—The autopsy was confined to an examination of skull, brain, and abdominal viscera. The body was in a

state of extreme emaciation ; all muscles relaxed. The skull was thick, and skull cap unusually heavy. Outer and inner surfaces smooth and showed no unusual appearances or impressions. Skull symmetrical ; left frontal fossa a trifle deeper than right ; large fontanelle very nearly ossified. A large organized clot was found in the superior longitudinal sinus ; there was some thickening of the dura to either side of the sinus, some slight adhesion over the upper portion of the precentral and over the left temporal convolution, but even here and over the entire surface of the brain the pia could be easily removed without injuring the parts below. There was an œdema of the entire convexity ; unusual pallor of the convolutions ; no marked increase of the fluid in the lateral ventricles. Freed of its dura, the brain weighed exactly two pounds (one thousand grams). Blood-vessels appeared normal and had normal distribution. I may state at once that the cortex was hard to the touch, and that the knife grated in removing a small portion of the cortex for immediate examination. This grating was due to small calcified plates. On superficial inspection, the great breadth of the fissures, the corresponding narrowness of the convolutions, and the unusual exposure of the left island of Reil were very apparent. A detailed examination of the larger ganglia, of the pons, medullary, etc., will be made and will be reported upon later on.

The spleen was enlarged and the liver hard, but no evidences of hereditary syphilis.

EXAMINATION OF BRAIN.

The brain was immersed at once into Müller's fluid, and as soon as hardened the brain surfaces were photographed.¹ By comparison with the paper² which our retiring president read last year, you will recognize certain departures

¹ I am indebted to Mr. O. G. Mason for the original photographs, but one of which is reproduced in this article ; all four photographs were exhibited at the meeting of the Association.

² C. K. Mills, Presidential Address (*JOURNAL OF NERVOUS AND MENTAL DISEASE*, vol. xiii., 1886).

from normal fissuration which are indicative of inferior brain development.

Examination of Brain Surfaces.

Left hemisphere, outer surface. (Plate I.)

The most striking features are the great depth of all fissures, and the comparative simplicity of fissuration, particularly in the frontal lobes; the great exposure of the island of Reil due to the retraction and narrowness of the surrounding convolutions. The central fissure (*c*) is bifurcated and is clearly confluent with the Sylvian fissure which is broad and long. The first temporal fissure (*t. 1*)—supertemporal, Wilder—would be continuous high up into the parietal region, but for a slight bridging convolution. The parieto-occipital fissure is unusually distinct and in the occipital lobe the three fissures are easily traced. In the frontal lobe, the first and second frontal fissures are well marked, while the second forms the long branch of a zygial formation according to Wilder. The convolutions appear alternately narrowed and broadened; this is particularly true of the first temporal and precentral convolutions. The gyrus angularis is scantily developed.

The mesial surface of left hemisphere exhibits the confluence of the parieto-occipital, the calcarine and hippocampal fissures. The collateral fissure of Wilder well marked. The calloso-marginal fissure well defined though shallow. The præcuneus massive, the cuneus of normal size.

Right hemisphere—*outer surface*.

Here the conditions approach much more nearly to the normal. The island of Reil is scarcely exposed; the fissure of Sylvius of normal breadth and length; the central fissure is confluent with the fissure of Sylvius. The first temporal convolution is *continuous* into the parietal region, and there is a distinct though very narrow angular gyrus. Wilder's interparietal fissure is distinct; in both the occi-

pital and frontal lobes, three typical fissures can be made out; there is an undoubted medifrontal (Wilder) fissure which could not be traced on left side. The parieto-occipital does not form as distinct an indentation as on left outer surface.

Median surface.—The parieto-occipital, calcarine, and hippocampal fissures are confluent; the collateral fissures well defined; the entire mesial surface is divided into small blocks by numerous secondary fissures. Cuneus and præcuneus of normal development.

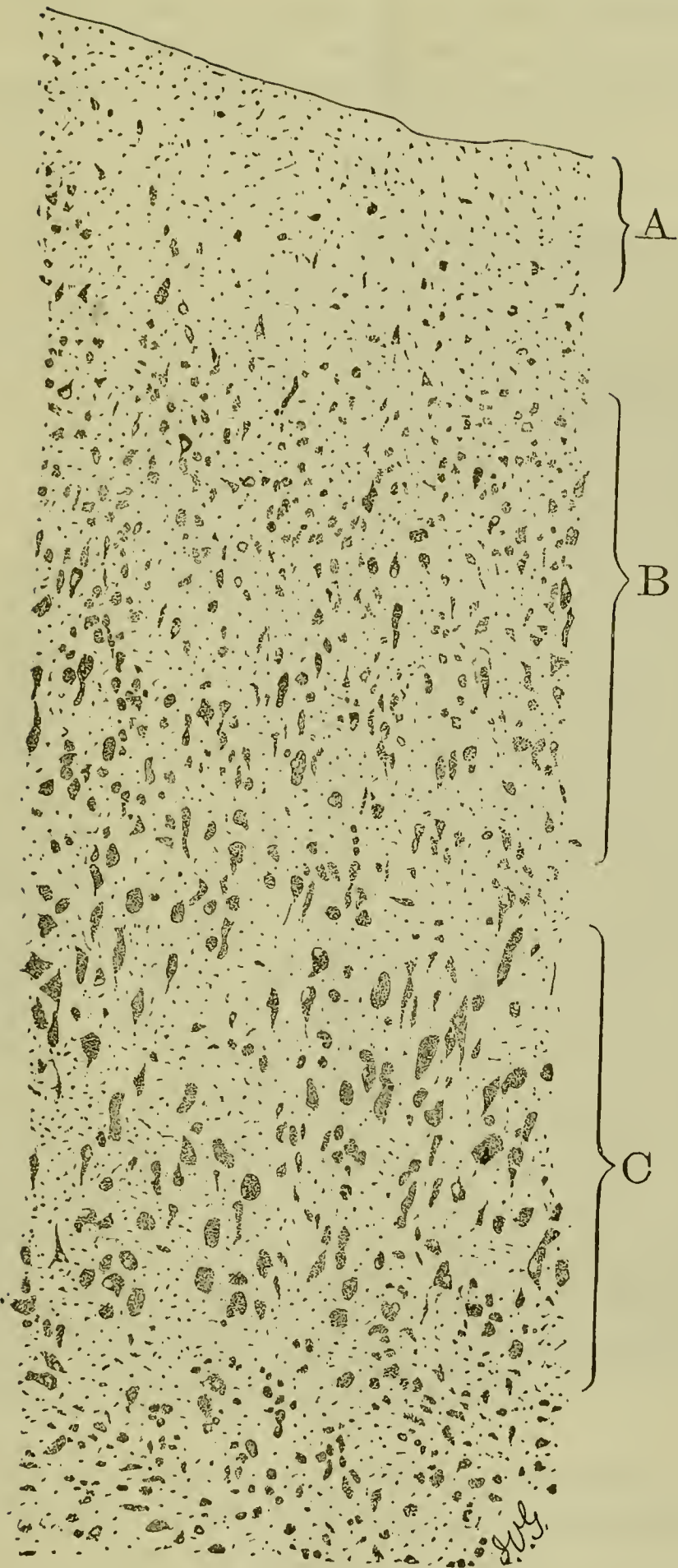
Microscopical Examination of the Cortex.

The brain surfaces, after they had been thoroughly hardened in Müller's fluid, were cut up into small blocks for histological examination. Sections from the frontal lobes, the motor zones, the base of the third frontal convolution, from the first temporal convolution, and from the occipital apex of both hemispheres have been examined. The cuneus was unfortunately too brittle to permit of section cutting. From the portions thus far examined, it is fair to infer that the changes to be described affect equally every part of the brain surface. The plates herein given represent the changes as seen in sections from the first temporal convolution of the left side. These specimens were stained according to the acid fuchsin method, others were stained with Weigert's two hæmatoxylin methods, and with ammoniacal carmine. You will note that the cellular elements exhibit the same changes, whatever staining method we employed. On the drawing, most carefully made by Dr. Van Gieson, and in these specimens¹ the following conditions may be noted.

We are able to distinguish the external barren layer, the layer of small pyramidal cells, the layer of the large pyramids, and perhaps a trace of Meynert's fourth granular layer. Examining these sections, very marked.

¹ Demonstrated at the meeting.

PLATE II.



changes will be observed in the structure of the small and large pyramid cells. In my search through the entire brain I have not come across more than half a dozen, if as many, pyramid cells of anything like normal appearance.

PLATE III.



The fewest large and small pyramid cells show well-defined processes. The contours are rounded, and the cell substance exhibits every possible change of its protoplasmic substance. In some there are a distinct nucleus and nucleolus, surrounded by a detritus-like mass; in many

the nucleus and nucleolus are entirely wanting. All these varied changes can be studied best with the acid fuchsin method; in Weigert preparations, the whole pathological cell mass takes up the stain deeply, and it is not always easy to distinguish the nucleus and cell-body. Glancing through the sections, you will also observe that a few of the cells turn their apices downward instead of upward, thus exhibiting a change to which Brückner refers as occurring in his case of tuberos sclerosi and to which no pathological significance is to be attached.

Plate III. exhibits these changes under a very much higher power. In some cells a partly normal and a partly pathological character of the cell-body is visible. In the neuroglia, I have not been able to prove any changes; there is certainly no sclerosis visible in any part I have examined. The white fibres have not undergone morbid changes, but on Weigert specimens they cannot be traced as far towards the periphery as in the normal cortex; the transverse fibres in the outer barren layer could not be made out. There is no evidence whatever of any previous encephalitic process. No infiltration around the blood-vessels; in fact no changes in any of the blood-vessels of the cortex. At the meeting, doubts were expressed whether there was not a paucity of blood-vessels. I have paid special attention to this point, and am now convinced, after examining a very large number of sections from every part of the cortex, that these capillary vessels are of normal calibre and as numerous as in corresponding sections of the normal brain. Nor is there any proliferation of the nuclei of these cells in the walls of the blood-vessels. We have then a simple change affecting the cells and possibly the white fibres only, and the question remains to be decided whether there is mere arrest of development, or an arrest of development the result of a previous inflammatory process. There is nothing in support of the latter proposition, and everything in favor of the former.

I cannot find any evidence of distinct degenerative changes in the cells, and it would seem to me that, if the:

process were one that had set in after the cells had already matured, we should find some, and many more cells than we actually do, exhibiting a more complete formation than any to be found on the specimens before you. You will note also that there were no gross changes such as are frequently held responsible for insufficient development: there is no evidence of hydrocephalus internus, of a general or a multiple tuberous sclerosis; no traces of a preceding encephalitis.

We have here an agenetic condition pure and simple, affecting the highest nerve elements. As to the cause of this agenetic condition, I am not willing to speculate. I repeat that syphilis is excluded, at least not proved, that there is strong hereditary predisposition to mental troubles, and that there is the etiological factor of traumatism in the case. As the foetal circulation is easily affected by the slightest disturbances, and the proper nutrition of the most highly differentiated organ of the body may in this way have become impaired, we cannot afford to overlook the factor of traumatism.

EXPLANATION TO PLATES I., II., AND III.

PLATE I.

Outer aspect of surface of left hemisphere showing the exposure of the island of Reil, and great breadth of fissure of Sylvius.

X denotes region from which first block of cortical tissue had been removed for histological examination.

C, central fissure or fissure of Rolando.

prc, precentral fissure.

i. p, interparietal fissure.

oc, occipital; parieto-occipital fissure.

*t*¹, *t*², first and second temporal fissures.

*f*¹, *f*², first and second frontal fissures.

Other explanations in text.

PLATE II.

× 70 diameters.

Section from first temporal convolution; specimen stained with acid fuchsin; drawing made with especial reference to changes in the cells.

Divisions A, B, C, correspond about to layers of superficial neuroglia, of small pyramid cells and of large pyramid cells. Below C is fourth granular layer (Meynert).

It will be noted that with this low magnifying power, the changed appear-

ance of the pyramid cells can be made out. The contours of the cells are altered, the pyramidal shape is often widely departed from; the cell body is altered, and occasionally shows distinct lacunæ.

PLATE III.

× 500 diameters.

Section of first temporal convolution, representing a portion of division C under much higher magnifying power. Pyramidal cells have lost their normal shape. The cell body has a homogeneous but altered appearance; nuclei either absent or distorted; smaller cells and neuroglia cells with distinct nuclei; section of capillary vessels normal, in the upper right hand corner a distorted cell mass with pericellular space around.

